Sensory loss in syringomyelia: not necessarily dissociated

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Summary
The pattern of sensory loss was assessed in 210 cases of syringomyelia. Dissociated sensory loss occurred in 49% of cases, indicating that its occurrence is not a necessary finding for the diagnosis of syringomyelia. Syringomyelia should be considered in the differential diagnosis of all cases of spinal cord disease.

Introduction
Syringomyelia is a well known disorder of the spinal cord. It is categorized pathologically by the presence of longitudinal cavities within the spinal cord, most commonly the lower cervical and upper dorsal regions[1]. The textbook clinical presentation is with wasting and weakness in the upper limbs, a spastic paraparesis and often pain in the head, neck or limbs. However, the clinical feature most often emphasized, and remembered by most medical students, is the dissociated sensory loss[2-7]. One textbook[4] states that segmental dissociated sensory loss, together with amyotrophy of the upper limbs are fundamental, and that the clinical diagnosis of syringomyelia can hardly be made without them. The classic description of dissociated sensory loss is of loss of pain and temperature sensation, in a cape or half-cape distribution, together with preservation of light touch; vibration and joint position sense loss are taken as evidence of dorsal column involvement[3,4,8]. The explanation given for this distribution of clinical signs is that the syrinx cavity disrupts the decussating sensory fibres in the grey and white matter passing from the dorsal horn to the opposite lateral spino-thalamic tract[3,4].

Although atypical presentations have long been recognized[2], the advent of MRI scanning has highlighted the presence of syringomyelia in cases without the characteristic clinical features[9]. Williams has previously written about the typical presentation of syringomyelia and has also commented on the variability of clinical presentation[6,10]. The present study was intended to focus on sensory loss in syringomyelia, in particular to quantify the frequency of dissociated sensory loss in syringomyelia, in order to determine its reliability as a clinical predictor of the diagnosis.

Methods and results
The study is based on a database of 606 cases of syringomyelia and related disorders, which has been collected by the senior author (BW) over a number of years. In conjunction with other aspects of syringomyelia, the database records the findings on sensory examination in each case. Dissociated sensory loss

was defined as loss of pain or temperature sensation with preservation of other sensory modalities, including light touch, vibration and joint position sense. Posterior column sensory loss was considered to be present if there was impairment of joint position sense or two-point discrimination. A further group was designated to include cases in which both pain or temperature and dorsal column sensation were impaired.

In order for the cases to be as homogeneous as possible, it was decided to limit the inclusion criteria to cases with certain syringomyelia associated with hind brain herniation; other causes of syringomyelia were deliberately excluded. The diagnosis of syringomyelia was confirmed at operation in all but 10 cases; in these cases the diagnosis was based on appropriate neuro-radiological imaging. These inclusion criteria were fulfilled in 214 cases, four cases were discarded because of incomplete information, leaving 210 cases on whom the analysis was undertaken.

Sensory examination was carried out by the senior author (BW) in all cases. The following sensory modalities were tested; light touch, pin prick, vibration, joint position sense and two-point discrimination. The findings on sensory examination were as follows: dissociated sensory loss occurred in 102/210 (49%) cases; posterior column loss alone occurred in 17/210 (8%) cases and both posterior column and lateral spinothalamic loss occurred in 36/210 (17%) cases. In 55/210 (26%) cases no sensory loss was detected. The findings may be represented as a Venn diagram (Figure 1).

It was noted that some patients were able to respond clearly to the clinical sensory examination and, for example, describe loss of pain sensation, with preservation of other sensory modalities. However, other patients, often the intelligent or introspective, found it difficult to describe their sensory loss in a clear-cut 'yes or no' way in response to each sensory modality; phrases such as 'a sort of numbness', or 'it's as if I have thin gloves on' were often used. In such clinical circumstances it is possible that in the presence of a major sensory deficit, a minor deficit might be concealed. If the sensory modalities had been tested using more precise psychophysical techniques, then we might have found that isolated dissociated or posterior column sensory loss rarely occurred.

Discussion
This study of a large population of syringomyelia has shown that the classical sensory feature of dissociated sensory loss only occurs in around half of the cases. About a quarter of cases have no sensory loss and a significant minority of cases have posterior column loss only.
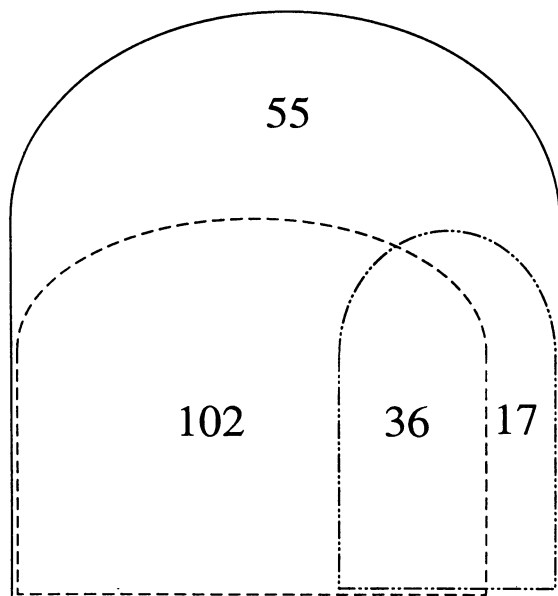
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Figure 1. Venn diagram to illustrate the disposition of the two main classes of sensory loss. ———— Pain and temperature loss; —··— proprioceptive and touch

The pattern of sensory loss in syringomyelia can be considered in terms of the pathology. The traditional view, as stated above, is that the dissociated loss is produced by interruption of the decussating spinothalamic fibres in the anterior commissure. This hypothesis assumes that the syrinx is usually placed in the region of the central canal. However, pathological studies have shown that syrinx location within the spinal cord is variable; a syrinx often occurs at other sites, including the dorsolateral portion of the cord[1,2,11]. Because of the structure of the ventral fissure and the dorsal columns, it is common for the cavity to be predominantly unilateral (Figure 2). It seems likely that the destruction of the posterior horn region may specifically disrupt the substantia gelatinosa, because of its structural properties, more frequently than the more medial cell relays passing
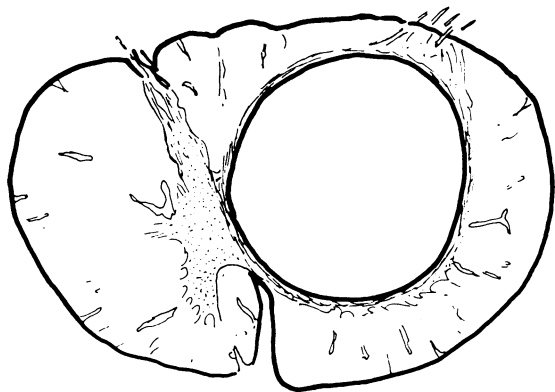

Figure 2. A moderate-sized syrinx is shown. The syrinx is unilateral and is causing severe distortion of the dorsal horn region

to the posterior columns. Minor variations in the destruction of the posterior horn region could account for the diversity of sensory signs which have been detected. It could also explain why dissociated sensory loss is frequently unilateral.

An additional factor may be the specificity of function within the anterolateral and dorsal column pathways. It has been suggested that two-point discrimination and joint position sense are not specific to the dorsal columns[12]. However, a study of sensation in cases with anterolateral cordotomy found loss of two-point discrimination or joint position sense occurred only in a small minority of cases[13].

In conclusion, many cases of syringomyelia will be missed if the diagnosis is only considered in cases with dissociated sensory loss. Williams has stated elsewhere that motor signs in hind brain related syringomyelia occur in up to 70% cases and that wasting is usually a late manifestation[10]. Therefore, we suggest that syringomyelia be considered in the differential diagnosis of all patients with clinical features suggestive of a spinal cord lesion.

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